

# Diagnostic and surgical management of patients with aneurysms of the thoracic aorta with various causes

## *Echocardiography and contrast enhanced computed tomography in prophylactic replacement of the ascending aorta*

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**SUMMARY** Sixty eight patients with aneurysms of the thoracic aorta were studied. Forty one had aortic dissection, 24 had dilatation only, and three had transverse aortic rupture. Sixteen had Marfan's syndrome; 17 had hypertension; and in eight there were other causal factors. In 17 the cause of the aneurysm was unknown. Histological examination did not help to establish the cause of aneurysm. Echocardiography failed to detect dissection of the ascending aorta in four (21%) out of 19 cases studied.

The mortality rate in the whole series was 26%. Early (operative and hospital) and late deaths occurred in 20% and 6% of patients respectively. The early mortality rate was 40% in the 24 emergency cases of dissection of the ascending aorta, 9% in patients operated on for dilatation of the ascending and transverse aorta without dissection, and 8% in patients with chronic dissection of the ascending aorta who had elective operation. Early and late mortality rates were no higher in patients with Marfan's disease than in any of the other groups.

It is suggested that contrast enhanced computed tomography should be performed in all patients with pronounced aortic root dilatation and in patients with Marfan's disease with symptoms which suggest dissection, even if they have only slight aortic root dilatation. Preventive replacement of the ascending aorta should be considered in more patients to reduce the number of emergency operations, in which the mortality rate is high. There is no definite limit of aortic root dilatation above which preventive replacement of the ascending aorta should be routinely considered.

Angiography and echocardiography have greatly increased the accuracy of diagnosis of aneurysms of the thoracic aorta. Surgery is now the preferred treatment for aneurysms, especially for those of the ascending aorta. New methods of investigation will improve knowledge of the anatomy of these lesions and may lead to improvement in their management.

We report the results of surgery in 68 patients

with aortic aneurysm, together with the value of echocardiography and contrast enhanced computed tomography in managing these patients.

### **Patients and methods**

We studied 68 consecutive patients operated on between January 1979 and December 1982. There were 54 men, mean (SD) age 47.6 (13.3) years (range 14-69) and 14 women, mean age 42.6 (13.3) years (range 21-65). Twenty seven had acute symptoms and 41 had chronic symptoms which suggested aortic lesions.

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## SKELETAL AND OPHTHALMOLOGICAL EVALUATIONS

Skeletal abnormalities were assessed by the metacarpal index (normal values under 8.4) and the difference between height and arm span (normal values <7.6 cm).<sup>1</sup> Patients with subluxation of the lenses and abnormalities of the anterior chamber angle were classified as probable (+ +) or possible (+) cases of Marfan's syndrome.<sup>2</sup>

## CARDIOVASCULAR EVALUATION

**Echocardiography**—M mode and cross sectional echocardiograms were recorded before operation in 36 patients who were examined for the presence of the following conditions: aortic root dilatation by criteria suggested by Brown *et al*.<sup>3</sup>; aortic root dissection, and mitral valve prolapse. Serial studies were possible preoperatively in 13 patients with chronic lesions of the aorta (with at least three examinations at intervals of six months) and in 10 of them contrast enhanced computed tomography was performed. Increases of more than 10% of the aortic root diameter measurement were regarded as important.

**Angiography**—Aortography and left ventricular angiography with selective coronary arteriography were performed in all patients with dilatation of the ascending aorta, and in those in whom dissection of the ascending aorta was suspected. Aortography alone was performed in patients with dissection of the descending aorta. We defined aortic dissections as type I, II, III, according to De Bakey's classification. Severity of aortic valve and mitral valve regurgitation was assessed by angiography on a scale of 1 to 4. Grades 3 and 4 indicated severe regurgitation. Angiography was performed after operation in eight cases in which dissection extended from the ascending aorta into the arch and beyond (type I); in two with dissection only of the ascending aorta (type II); and in three with dissection of the descending tract of the thoracic aorta (type III).

**Contrast enhanced computed tomography** was performed before operation in 10 patients in whom dilatation of the ascending aorta had been diagnosed by echocardiography and after operation in 17 patients (11 operated on for type I dissections, three for type II, two for type III, and one for rupture of the transverse aorta). Both angiography and contrast enhanced computed tomography were performed postoperatively in five patients, four of whom had been operated on for type I dissections. Contrast enhanced computed tomography was performed with a Siemens Somatom 2 at a scan speed of 3.5 seconds after multiple rapid peripheral intravenous

injections of 25–50 ml of Urografin (Schering) up to a maximum of 200 ml.

## SURGICAL TREATMENT

In the absence of contraindications, medical treatment (antihypertensive and negative inotropic agents) was given in the first hours after admission to hospital and during angiography in an attempt to stabilise the patient's condition. An operation was considered immediately if medical treatment was contraindicated or when, despite medical treatment, there was rapid deterioration of the patient's haemodynamic state or persistent pain suggestive of progressive dissection.

For aneurysms of the ascending aorta and in type I and type II dissections the tract of affected aorta was replaced through a median sternotomy by a low porosity Dacron graft with or without reimplantation of the coronary arteries. In patients with severe aortic regurgitation and aneurysms or dissections, a composite graft with a valve prosthesis (Sorin) was used except in three patients with dilatation of ascending aorta and severe aortic regurgitation in whom a valve prosthesis and a vascular prosthesis were implanted. In cases of dissection the false channel was obliterated and the two layers affected by the dissecting process were cuffed proximally and distally with Teflon.

The dilatations and dissections of the descending thoracic aorta were approached through a posterolateral thoracotomy, with cannulation of the left femoral vein and artery for partial bypass to perfuse the legs during cross clamping.

For cerebral protection we used moderate hypothermia (25°C) during operations on aneurysms and type II and III dissections and profound hypothermia (15°C) in type I dissections.<sup>4</sup> In profound hypothermia cooling and rewarming times increased with the patient's body surface area. A cardioplegic solution was introduced directly into the coronary ostia and pericardial ice slush was used to produce local cardiac cooling.

The sections of aorta removed at operation were examined to determine the severity of cystic medial necrosis, elastin fragmentation, fibrosis, and medionecrosis, and compared with those of 30 normotensive subjects of similar age.

Late follow up of survivors ranged from 24 months to six years (mean 47 months).

## DEFINITIONS

Patients were described as having Marfan's syndrome if they had typical abnormalities of at least two of the three organ systems evaluated; they were described as having formes frustes if, together with the aortic abnormalities, one only of the two assess-

Table 1 Clinical diagnosis of the thoracic aortic lesions

|                                  | Male patients | Female patients | Total, mean (SD) age, and age range (yr) |
|----------------------------------|---------------|-----------------|--|
| No of patients                   | 54            | 14              | 68                                       |
| Acute dissection                 | 21            | 3               | 24<br>44.7 (12.1)<br>19-65               |
| Chronic dissection               | 15            | 2               | 17<br>54.3 (9.8)<br>27-68                |
| Type I                           | 20            | 4               | 24<br>45.1 (11.4)<br>21-65               |
| Type II                          | 10            |                 | 10<br>52.3 (13.6)<br>19-68               |
| Type III                         | 6             | 1               | 7<br>56.0 (8.0)<br>42-64                 |
| Rupture of the transverse aorta  | 1             | 2               | 3<br>14-40                               |
| Aneurysm without dissection      | 17            | 7               | 24<br>45.7 (13.0)<br>29-69               |
| Aneurysm of the ascending aorta  | 13            | 2               | 15<br>44.7 (12.7)<br>29-65               |
| Aneurysm of the transverse aorta | 2             | 5               | 7<br>43.3 (12.6)<br>29-62                |
| Aneurysm of the descending aorta | 2             |                 |  |

ments for skeletal evaluation was abnormal or both showed values at the upper limits, or if there were moderate alterations of the anterior ocular chamber angle.

Patients were deemed to have hypertension if there was a history of drug treatment for diastolic hypertension or if their diastolic pressure was higher than 99 mm Hg.

Trauma, aortic valve stenosis, aortic valve prosthesis, bicuspid aortic valve, and coarctation of the aorta were regarded as other causal factors; but a possible association with Marfan's syndrome, or hypertension was always considered.<sup>5-7</sup>

When none of the conditions described above was present the aneurysm was described as being of unknown cause. We subdivided patients into four groups on the basis of these definitions.

## Results

### CARDIOVASCULAR EVALUATION

Table 1 shows the clinical diagnoses of the 68 patients.

### AETIOLOGICAL AND ANATOMICAL FINDINGS

#### *Lesions in patients with connective tissue disease*

Skeletal, ophthalmological, and cardiovascular examinations led to the diagnosis of connective tissue disease in 16 patients. Ten had typical Marfan's syndrome and six had formes frustes of Marfan's syndrome. Type I dissections were seen in five patients with typical features of Marfan's syndrome and in three with formes frustes (Table 2). In four other cases with typical features and in two with formes frustes there was dilatation of the ascending aorta (Table 3); in one woman with formes fruste and history of a recent trauma, there was dilatation of the transverse aorta (case 44). Emergency operations were performed in five subjects with type I dissections and in one boy with rupture of the transverse aorta. All six had features typical of Marfan's syndrome. In the boy with rupture of the transverse aorta, the metacarpal index was 9.1, there were ocular abnormalities, and mitral valve prolapse (without dilatation of the aortic root) was present. Mitral valve prolapse was observed in three of the 16 patients. They all had aortic dilatation, except the boy with rupture of the transverse aorta and the woman with dilatation of the transverse aorta.

#### *Lesions in patients with hypertension*

Aortic dissection was seen in 19 hypertensive patients; type I in eight, type II in five, and type III in six (Table 2). Dilatation of the ascending aorta was seen in four patients, of the transverse aorta in one, and of the descending aorta in two (Table 3). Ten had emergency operations.

#### *Lesions in patients with aneurysms and other causal factors*

Traumatic rupture of the transverse aorta was seen in two women, one with diastolic hypertension; traumatic dilatation of the unruptured aorta was seen in two other women (cases 42 and 44), one of whom had an abnormal metacarpal index and a mitral valve prolapse (Table 3). A congenital bicuspid aortic valve was seen in one man with type II chronic dissection and hypertension (case 28) and in one man with dilatation of the ascending aorta and Marfan's syndrome (case 49). There was coarctation of the aorta in a boy with type II acute dissection (case 4) and in a woman with dilatation of the transverse aorta (case 45). In one man (case 37) with aortic valve stenosis and type II chronic dissection there was an abnormal height-arm span difference; in three other men who had had an aortic valve replaced, there were type I dissections in two (cases 15 and 35) and dilatation of only the ascending aorta in one (case 51).

Table 2 *Skeletal, ophthalmological, and cardiovascular examination in patients with acute or chronic aortic dissection\**

| Case No                    | Sex | Age (yr) | Evidence for inheritance of Marfan's syndrome | Height-span difference (cm) | Metacarpal index | Ocular evidence of Marfan's syndrome | Hypertension | Trauma | Echocardiography | Angiography                            | Remarks                  |
|----------------------------|-----|----------|---|-----------------------------|------------------|--------------------------------------|--------------|--------|------------------|--|--------------------------|
| <i>Acute dissection:</i>   |     |          |   |                             |                  |                                      |              |        |                  |  |                          |
| 1                          | F   | 33       | No  | -2                          | 8.4              | +                                    | No           | No     | Dilatation       | Type I                                 | Marfan's syndrom         |
| 2                          | F   | 41       |   |                             |                  |                                      |              | No     |                  | Type I, MVP                            | Unknown cause†           |
| 3                          | F   | 47       |   |                             |                  |                                      |              | No     | Dissection       | Type I                                 | Unknown cause†           |
| 4                          | M   | 19       |   |                             |                  |                                      |              | No     |                  | Type II, SAR, coarctation of the aorta | Other cause†             |
| 5                          | M   | 21       | Yes   | -6                          | 8.6              | ++                                   | No           | No     | Dissection       | Type I, SAR                            | Marfan's syndrom         |
| 6                          | M   | 30       | No  | -7.6                        | 8.7              | No                                   | No           | No     | Dissection       | Type I, SAR                            | Marfan's syndrom         |
| 7                          | M   | 30       | No  | -7.5                        | 9.0              | No                                   | No           | No     | Dissection       | Type I                                 | Marfan's syndrom         |
| 8                          | M   | 40       | No  | 3                           | 7.9              | No                                   | Yes          | No     |                  | Type I                                 | Hypertension             |
| 9                          | M   | 42       | No  | -2                          | 8.6              | No                                   | No           | No     | Dissection, MVP  | Type I, MVP                            | Marfan's syndrom         |
| 10                         | M   | 42       |   |                             |                  |                                      |              | No     | Dilatation       | Type I, SAR                            | Unknown cause†           |
| 11                         | M   | 42       |   |                             |                  |                                      | Yes          | No     | Dissection       | Type I, SAR                            | Hypertension†            |
| 12                         | M   | 42       |   |                             |                  |                                      | Yes          | No     |                  | Type III                               | Hypertension             |
| 13                         | M   | 42       | No  | -5                          | 7.8              | No                                   | No           | No     | Dissection       | Type I                                 | Unknown cause            |
| 14                         | M   | 47       |   |                             |                  |                                      |              | No     | Dissection       | Type I                                 | Unknown cause            |
| 15                         | M   | 47       |   |                             |                  |                                      |              | No     |                  | Type I, SAR                            | Other cause†             |
| 16                         | M   | 49       |   |                             |                  |                                      | Yes          | No     |                  |  | aortic valve replacement |
| 17                         | M   | 50       |   |                             |                  |                                      | Yes          | No     |                  | Type II                                | Hypertension             |
| 18                         | M   | 51       | No  | -11                         |                  | No                                   | No           | No     |                  | Type I                                 | Hypertension†            |
| 19                         | M   | 55       | No  | -6.5                        | 7.9              | No                                   | Yes          | No     | Dissection       | Type II, SAR                           | Unknown cause            |
| 20                         | M   | 56       | No  | -6                          | 7.8              | No                                   | Yes          | No     | Dissection       | Type I                                 | Hypertension             |
| 21                         | M   | 59       |   |                             |                  |                                      | Yes          | No     |                  | Type I                                 | Hypertension             |
| 22                         | M   | 61       |   |                             |                  |                                      | Yes          | No     | Dissection       | Type II                                | Hypertension†            |
| 23                         | M   | 62       |   |                             |                  |                                      | Yes          | No     |                  | Type I                                 | Hypertension†            |
| 24                         | M   | 65       |   |                             |                  |                                      | Yes          | No     |                  | Type III                               | Hypertension†            |
|                            |     |          |   |                             |                  |                                      |              | No     |                  | Type I                                 | Unknown cause†           |
| <i>Chronic dissection:</i> |     |          |   |                             |                  |                                      |              |        |                  |  |                          |
| 25                         | F   | 52       |   |                             |                  |                                      | Yes          | No     |                  | Type I                                 | Hypertension             |
| 26                         | F   | 62       | No  | -2                          | 7.2              | No                                   | Yes          | No     |                  | Type III                               | Hypertension             |
| 27                         | M   | 27       | No  | -8                          | 11               | No                                   | No           | No     | Dilatation       | Type I, SAR                            | Marfan's syndrom         |
| 28                         | M   | 46       | No  | -5                          | 7.2              | No                                   | Yes          | No     |                  | Type II, bicuspid aortic valve         | Hypertension             |
| 29                         | M   | 46       | No  | 1                           | 8.8              | No                                   | No           | No     | Dissection       | Type I                                 | Marfan's syndrom         |
| 30                         | M   | 47       | No  | -7.5                        | 8.8              | No                                   | No           | No     | Dilatation       | Type I, SAR                            | Marfan's syndrom         |
| 31                         | M   | 50       |   |                             |                  |                                      | Yes          | No     |                  | Type III                               | Hypertension             |
| 32                         | M   | 51       |   | -1                          | 7.9              |                                      | No           | No     |                  | Type II, SAR                           | Unknown cause            |
| 33                         | M   | 53       | No  | 5                           |                  | No                                   | Yes          | No     |                  | Type III                               | Hypertension             |
| 34                         | M   | 56       |   |                             |                  |                                      | No           | No     |                  | Type II                                | Unknown cause            |
| 35                         | M   | 56       |   |                             |                  |                                      | No           | No     | Dissection       | Type I                                 | Other cause†             |
|                            |     |          |   |                             |                  |                                      |              |        |                  |  | aortic valve replacement |
| 36                         | M   | 59       | No  | -5.5                        | 8.15             | No                                   | No           | No     |                  | Type III                               | Unknown cause            |
| 37                         | M   | 59       | No  | -8                          | 7.85             | No                                   | No           | No     |                  | Type II                                | Other cause              |
|                            |     |          |   |                             |                  |                                      |              |        |                  |  | stenosis of aortic valve |
| 38                         | M   | 63       | No  | -6                          |                  | No                                   | Yes          | No     | Dissection       | Type I                                 | Hypertension             |
| 39                         | M   | 64       |   |                             |                  |                                      | Yes          |        |                  | Type III                               | Hypertension†            |
| 40                         | M   | 65       |   |                             |                  |                                      | Yes          |        |                  | Type II, SAR                           | Hypertension             |
| 41                         | M   | 68       | No  |                             |                  | No                                   | Yes          | No     | Dissection       | Type II                                | Hypertension             |

\*Three cases, including a boy with Marfan's syndrome, with rupture of the transverse aorta are not presented in the Table.

Cases 1, 9, and 29 have formes frustes of Marfan's syndrome.

Ocular evidence of Marfan's syndrome: ++ probable, + possible.

†Early (operative and hospital) death; ‡late death.

MVP, mitral valve prolapse; SAR, severe aortic regurgitation (slight aortic regurgitation, if present, is not mentioned); dilatation, aortic root dilatation; dissection, aortic dissection.

#### *Lesions in patients with aneurysms of unknown cause*

Acute dissection of the ascending aorta was seen in seven patients and chronic dissection of the ascending aorta in three (Table 2). There was dilatation of the ascending aorta in three cases and dilatation of the transverse aorta in another three

(Table 3). Mitral valve prolapse was observed in one woman at left ventricular angiography (case 2).

#### HISTOLOGICAL FINDINGS

The severity of cystic medial necrosis (graded as 1 to 4, according to the amount of basophilic ground

ble 3 Skeletal, ophthalmological, and cardiovascular evaluation in patients with aneurysm without dissection

| Case | Sex | Age (yr) | Evidence for inheritance of Marfan's syndrome | Height-span difference (cm) | Metacarpal index | Ocular evidence of Marfan's syndrome | Hypertension | Trauma | Echocardiography | Angiography               | Remarks                              |
|------|-----|----------|---|-----------------------------|------------------|--------------------------------------|--------------|--------|------------------|---------------------------|--------------------------------------|
| 2    | F   | 29       |   |                             |                  |                                      |              | Yes    |                  | TAA                       | Other cause                          |
| 3    | F   | 32       |   |                             |                  |                                      | No           | No     |                  | TAA                       | Unknown cause                        |
| 4    | F   | 36       | No  | -2                          | 9.5              | No                                   | No           | Yes    | MVP              | TAA, MVP                  | Marfan's syndrome and trauma         |
| 5    | F   | 38       |   |                             |                  |                                      | No           | No     |                  | TAA, coarctation of aorta | Other cause†                         |
| 5    | F   | 39       |   |                             |                  |                                      | No           | No     |                  | TAA                       | Unknown cause                        |
| 7    | F   | 62       | No  | -3                          | 7.4              | No                                   | Yes          | No     |                  | TAA                       | Hypertension                         |
| 8    | F   | 65       |   |                             |                  |                                      | Yes          | No     |                  | AAA, SAR                  | Hypertension†                        |
| 9    | M   | 29       | Yes   | -6                          | 7.3              | ++                                   | No           | No     | Dilatation       | AAA, SAR, bicuspid valve  | Marfan's syndrome                    |
| 10   | M   | 31       | No  | -6                          | 8.1              | No                                   | No           | No     | Dilatation       | AAA, SAR                  | Marfan's syndrome                    |
| 11   | M   | 32       | No  | 3                           | 7.9              | No                                   | No           | No     | Dilatation       | AAA, SAR                  | Other cause aortic valve replacement |
| 2    | M   | 33       | Yes   | -7.5                        | 8.8              | ++                                   | No           | No     | Dilatation       | AAA, SAR                  | Marfan's syndrome                    |
| 3    | M   | 34       | No  | -8                          | 8.4              | No                                   | No           | No     | Dilatation       | AAA, SAR                  | Marfan's syndrome                    |
| 4    | M   | 41       | No  | -5                          | 8.4              | ++                                   | No           | No     | Dilatation       | AAA, SAR                  | Marfan's syndrome                    |
| 5    | M   | 41       | No  | -4                          | 8.4              | No                                   | No           | No     | Dilatation       | AAA, SAR                  | Marfan's syndrome                    |
| 6    | M   | 42       |   |                             |                  |                                      | No           | No     | Dilatation       | AAA, SAR                  | Unknown cause†                       |
| 7    | M   | 45       |   |                             |                  |                                      | Yes          | No     | Dissection       | AAA, SAR                  | Hypertension                         |
| 8    | M   | 51       |   |                             |                  |                                      | No           | Yes    | Dilatation       | AAA                       | Other cause                          |
| 9    | M   | 54       | No  | -1                          | 7.0              | No                                   | Yes          | No     | Dilatation       | DAA, AAA                  | Hypertension                         |
| 10   | M   | 54       |   |                             |                  |                                      | Yes          | No     | Dilatation       | AAA                       | Hypertension                         |
| 11   | M   | 55       | No  | -7                          | 7.1              | No                                   | No           | No     |                  | TAA                       | Unknown cause                        |
| 12   | M   | 58       |   |                             |                  |                                      | No           | No     | Dilatation       | AAA                       | Unknown cause†                       |
| 13   | M   | 63       | No  | -7                          | 7.5              | No                                   | No           | No     | Dilatation       | AAA, SAR                  | Unknown cause                        |
| 14   | M   | 63       |   |                             |                  |                                      | Yes          | No     | Dilatation       | AAA                       | Hypertension                         |
| 15   | M   | 69       |   |                             |                  |                                      | Yes          | No     |                  | DAA                       | Hypertension                         |

cases 44, 50, and 55 had formes frustes of Marfan's syndrome.

Clear evidence of Marfan's syndrome: ++ probable.

†ly (operative and hospital) death; ‡late death.

Dilatation, aortic root dilatation; MVP, mitral valve prolapse; AAA, ascending aortic aneurysm; TAA, transverse aortic aneurysm; DAA, descending aortic aneurysm; SAR, severe aortic regurgitation.

substance and fragmentation of the elastic tissue) and the other histological findings were not significantly different in patients with aneurysm of unknown cause and in 30 normotensive subjects of similar age. There was no difference between patients with aneurysm and hypertension and the group of controls.

The degree of cystic medial necrosis in Marfan's syndrome was very variable and, in some cases, histological lesions were minimal.

#### ECHOCARDIOGRAPHIC FINDINGS

Echocardiography was performed in 19 consecutive patients in whom ascending aortic dissections were diagnosed by angiography (Table 2). Cross sectional echocardiography detected aortic root dissection in 15 patients and M mode echocardiography detected dissection in nine. In four patients M mode and cross sectional echocardiography did not show aortic root dissection (Fig. 1). Echocardiography was also performed in 16 patients in whom aneurysms without dissection (Table 3) were diagnosed by angiography. In 14 of these, dilatation of the aortic root was detected (Fig. 2) and in nine there were signs of aortic root dilatation together with aortic valve

regurgitation. In one patient (case 59), in whom aortic root diameter was moderately dilated, angiography showed a large aneurysm of the thoracic descending aorta that had been undetected by cross sectional echocardiography. In a woman with angio-

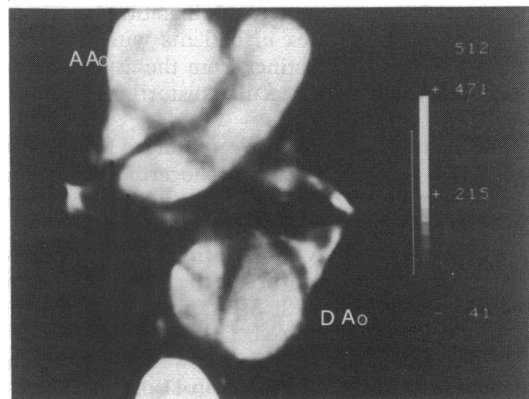


Fig. 1 Contrast enhanced computed tomograph in case 30, showing type I aortic dissection. This lesion was not detected by echocardiography. AAo, ascending aorta; DAo, descending aorta.

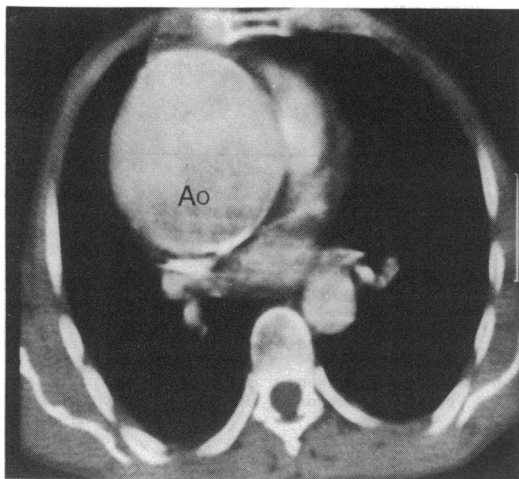


Fig. 2 A contrast enhanced computed tomograph in case 60 confirming the results of an echocardiogram which showed severe aneurysm of ascending aorta and excluded dissection. Aortic calcifications are evident. Ao, aorta.

graphic evidence of dilatation of the transverse aorta (case 44), echocardiography showed mitral valve prolapse but not the aortic dilatation. In a patient (case 57) dissection of the transverse aorta was incorrectly diagnosed (Fig. 3); however, in this subject ascending aortic aneurysm and aortic valve regurgitation were recognised by echocardiography.

In all patients aortic dilatation appeared to be symmetrical at M mode echocardiography and the precise location and extent of the aneurysm could not be detected. Identification of the aortic sinuses by cross sectional echocardiography enabled us to discern the site of the aneurysm or at least its starting point. We saw symmetrical dilatation beginning in the aortic sinus area in patients with Marfan's syndrome; this was distinct from the asymmetrical dilatation (often with saccular distortion) that was caused by other factors.

The diameter of the aortic root ranged between 50 and 86 mm by cross sectional echocardiography and measurements were slightly greater than those found at M mode echocardiography.

Echocardiography was performed only in one of the three cases with rupture of the transverse aorta. In this case—a boy with Marfan's syndrome—the rupture was not recognised.

Preoperative serial studies showed an important increase in aortic root diameter in 11 of the 13 cases. In the remaining two cases, in which contrast enhanced computed tomography diagnosed dissection, echocardiography did not show dissection or any increase of the aortic root dilatation.

## SURGERY

The operative and hospital mortality rate in patients with dissection from all causes was 29%, and late mortality was 5% (Table 4). The hospital mortality rate in patients with aortic dilatation without dissection was 8%; in those with ascending and transverse aortic dilatation it was 9%, and late mortality was also 9% in this group (Table 5).

Among patients with acute dissection, there were 10 (41%) operative and hospital deaths and one late death. Among patients with dissection who had elective surgery there were two (12%) hospital deaths (one (8%) in 12 cases of chronic ascending aortic dissection) and one late death at reoperation three months after the first operation. Among the three cases of rupture of the transverse aorta there were no hospital or late deaths.

There were six (31%) deaths among 19 patients with severe aortic regurgitation and aneurysms (8) or dissection (11) who were given a composite graft with a valve prosthesis and had reimplantation of the coronary arteries.

One of the three patients with aneurysms of the ascending aorta and severe aortic regurgitation (in whom a valve prosthesis and a vascular prosthesis were implanted) died in hospital (case 48). In patients with Marfan's syndrome and type I dissection there were no hospital deaths and one late death (Table 4). All seven patients with aortic dilatation without dissection survived (Table 5). In subjects with hypertension, there were six (31%) operative or hospital deaths, three in patients with type I dissections, one in a patient with type II dissection, and two in patients with type III dissection (Table 4). One hypertensive patient with dilatation of the ascending aorta without dissection died in

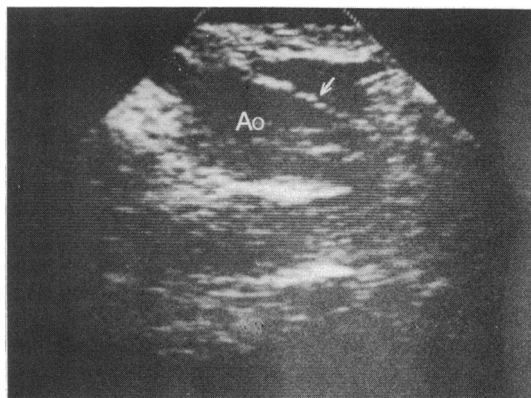


Fig. 3 An echocardiogram in case 57 suggesting the presence of dissection of transverse aorta. Dissection was excluded at angiography and at surgery. Arrow indicates the line responsible for the misinterpretation. Ao, aorta.

Table 4 Surgical results in cases of acute and chronic dissections according to cause

|   | No of patients | Operative deaths | Hospital deaths | Late deaths      |
|---|----------------|------------------|-----------------|------------------|
| <i>Acute aortic dissection:</i>                 | 24             | 5                | 5               | 1                |
| Connective tissue disease                       | 5              |                  |                 |                  |
| Hypertension                                    | 10             | 2                | 3               |                  |
| Other cause                                     | 2              | 1                |                 | 1 (at 4 months)  |
| Unknown cause                                   | 7              | 2                | 2               |                  |
| <i>Chronic aortic dissection:</i>               | 17             | 1                | 1               | 1                |
| Connective tissue disease                       | 3              |                  |                 | 1 (at 3 months)* |
| Hypertension                                    | 9              |                  | 1               |                  |
| Other cause                                     | 2              | 1                |                 |                  |
| Unknown cause                                   | 3              |                  |                 |                  |
| <i>Type I dissection (acute and chronic):</i>   | 24             | 5                | 3               | 2                |
| Connective tissue disease                       | 8              |                  |                 | 1                |
| Hypertension                                    | 8              | 2                | 1               |                  |
| Other cause                                     | 2              | 1                |                 | 1                |
| Unknown cause                                   | 6              | 2                | 2               |                  |
| <i>Type II dissection (acute and chronic):</i>  | 10             | 1                | 1               |                  |
| Connective tissue disease                       |                |                  |                 |                  |
| Hypertension                                    | 5              |                  | 1               |                  |
| Other cause                                     | 2              | 1                |                 |                  |
| Unknown cause                                   | 3              |                  |                 |                  |
| <i>Type III dissection (acute and chronic):</i> | 7              |                  | 2               |                  |
| Connective tissue disease                       |                |                  |                 |                  |
| Hypertension                                    | 6              |                  | 2               |                  |
| Other cause                                     |                |                  |                 |                  |
| Unknown cause                                   | 1              |                  |                 |                  |

\*This patient required reoperation.

hospital. There were no late deaths in patients with hypertension (Table 5).

Among patients with aneurysms with other causes and with dissection of the ascending aorta there were two operative deaths and one late death (Table 4). In patients in this group with dilatation and without dissection there was one late death (Table 5). In the group with aneurysms of unknown cause there were four (40%) hospital deaths, all in patients with type I dissection (Table 4); in the six cases with dilatation without dissection there was one hospital death and one late death (Table 5).

#### FOLLOW UP AND CAUSE OF DEATH

In the patients with aortic dissections the causes of hospital deaths were bleeding in six, septicaemia with renal failure in three, mediastinitis in one, and cerebral ischaemia with progressive stage IV coma due to recurrent dissection of the aortic arch in one. Bleeding and septicaemia caused two late deaths. In patients with aortic dilatation only, hospital deaths were the result of mediastinitis in one and low cardiac output in the other. There were two late deaths, one in a patient who died suddenly, presumably from arrhythmia, and the other from bleeding.

Table 5 Surgical results in cases of dilatation without dissection according to cause

|                                       | No of patients | Operative deaths | Hospital deaths | Late deaths      |
|---------------------------------------|----------------|------------------|-----------------|------------------|
| <i>Aneurysms of ascending aorta:</i>  | 15             | —                | 2               | 1                |
| Connective tissue disease             | 6              | —                | —               | —                |
| Hypertension                          | 4              | —                | 1               | —                |
| Other cause                           | 2              | —                | —               | —                |
| Unknown cause                         | 3              | —                | 1               | 1 (at 3 months)  |
| <i>Aneurysms of transverse aorta:</i> | 7              | —                | —               | 1                |
| Connective tissue disease             | 1              | —                | —               | —                |
| Hypertension                          | 1              | —                | —               | —                |
| Other cause                           | 2              | —                | —               | 1 (at 10 months) |
| Unknown cause                         | 3              | —                | —               | —                |
| <i>Aneurysms of descending aorta:</i> | 2              | —                | —               | —                |
| Connective tissue disease             | —              | —                | —               | —                |
| Hypertension                          | 2              | —                | —               | —                |
| Other cause                           | —              | —                | —               | —                |
| Unknown cause                         | —              | —                | —               | —                |

Table 6 Postoperative angiography and contrast enhanced computed tomography results in 24 of 35 survivors at operation for dissection

| Dissection | No of patients | Angiography |                   | Contrast enhanced computed tomography |                   |
|------------|----------------|-------------|-------------------|---------------------------------------|-------------------|
|            |                | Normal      | Distal dissection | Normal                                | Distal dissection |
| Type I     | 19             |             | 8                 | 4                                     | 7                 |
| Type II    | 9              | 1           | 1                 | 2                                     | 1                 |
| Type III   | 7              |             | 3                 | 2                                     |                   |

Both angiography and contrast enhanced computed tomography were performed in five patients, four of whom had type I dissection.

In patients undergoing postoperative angiography and contrast enhanced computed tomography or both (Table 6) distal dissections were common (16/24, 66%). In the five patients in whom both angiography and contrast enhanced computed tomography were performed the diagnoses by the two methods were the same (four had type I and one had type II dissections).

Reoperation was necessary in three patients, two for bleeding and one for abdominal ischaemia resulting from compression of the true lumen by the false one. One patient with Marfan's syndrome and type I dissection (case 27) who had reoperation for bleeding and distal dissection after three months died of persistent bleeding and cardiac tamponade; the patient with hypertension and type I dissection (case 38) who had reoperation for abdominal ischaemia one year later is alive and symptom free; the patient with a formes frustes of Marfan's syndrome and dilatation of the transverse aorta (case 44) who had reoperation for bleeding two days later is also alive and symptom free.

No case of paraplegia—which is a frequent complication of operating on descending thoracic aortic aneurysms—was observed in the present series. This absence of neurological sequelae underscores the safety and usefulness of the femoral vein-femoral artery bypass in treating descending thoracic aneurysms.

## Discussion

### CAUSE AND TYPE OF AORTIC LESION

In this series men outnumbered women whatever the cause of the aneurysms; patients with acute aortic dissections were younger than those with chronic aortic dissection, and patients with Marfan's syndrome and type I dissections were younger than patients with hypertension and type III dissections. Eight patients with type I dissections, seven with dilatation only, and one with rupture of an undilated transverse aorta, were deemed to have connective tissue disease. It is likely that the number of such patients would have been greater if more skeletal and ophthalmological examinations had been performed or if less restrictive criteria had been followed. No

patient with Marfan's syndrome had type III dissection or dilatation of only the descending aorta. All eight patients with type III dissection (six cases) and dilatation of the descending aorta (two cases) had hypertension.

### HISTOLOGICAL EXAMINATION

We expected that histological examination would be helpful in reaching a diagnosis in patients with aneurysms of unknown cause. In fact, it was of no help. On the basis of our results, and according to those reported by others (a) cystic medial necrosis (pooling of mucoid material); (b) elastin fragmentation (disruption of elastin lamellae); (c) fibrosis (increase in collagen at the expense of smooth muscle cells); and (d) medionecrosis (areas with apparent loss of nuclei) increase progressively with age independently of the presence of arterial hypertension.<sup>8,9</sup>

The variability of the degree of cystic medial necrosis in Marfan's syndrome accords with reports of minimal histological lesions in patients with Marfan's syndrome and aortic dissection.<sup>10</sup> Thus diagnosis of Marfan's syndrome can only be based on a thorough clinical examination to detect skeletal, ocular, and cardiac manifestations.

### SURGERY

The mortality rate in the whole series was 26%. Early (operative and hospital) and late deaths occurred in 20% and in 6% of patients respectively. Early deaths occurred in 40% of acute cases of ascending aortic dissection, in 9% of patients operated on for dilatation of ascending and transverse aorta without dissection, and in 8% of patients with chronic dissection of the ascending aorta who had elective surgery.

Our results with surgery of acute ascending aortic dissection in 15 of 22 patients in whom medical treatment was contraindicated compare favourably with the mortality rate in untreated patients.<sup>11,12</sup> There was one early postoperative death among seven patients with dissection of the ascending aorta in whom medical treatment had produced stabilisation of the clinical state. Our operative mortality in these cases compares favourably with the mortality rate in patients treated medically.<sup>11-13</sup>

Table 7 Contrast enhanced computed tomography (CT) and angiography in 10 patients with echocardiographic diagnosis of dilatation of the ascending aorta

| Case No | Echocardiography*  | CT scan*           | Angiography            |
|---------|--------------------|--------------------|------------------------|
| 27      | Dilatation         | Dissection         | Type I dissection, SAR |
| 30      | Dilatation         | Dissection         | Type I dissection, SAR |
| 50      | Dilatation (80 mm) | Dilatation (88 mm) | AAA, SAR               |
| 52      | Dilatation (68 mm) | Dilatation (70 mm) | AAA, SAR               |
| 53      | Dilatation (57 mm) | Dilatation (62 mm) | AAA, SAR               |
| 54      | Dilatation (51 mm) | Dilatation (53 mm) | AAA, SAR               |
| 55      | Dilatation (60 mm) | Dilatation (64 mm) | AAA, SAR               |
| 56      | Dilatation (50 mm) | Dilatation (55 mm) | AAA, SAR               |
| 60      | Dilatation (58 mm) | Dilatation (62 mm) | AAA                    |
| 64      | Dilatation (67 mm) | Dilatation (74 mm) | AAA                    |

\*Numbers in parentheses are aortic root diameter measurements.

Dilatation, aortic root dilatation; dissection, aortic root dissection; AAA, ascending aortic aneurysm; SAR, severe aortic regurgitation.

The chronic dissections of the ascending aorta of our series were asymptomatic (60%) or at a relatively stable stage (onset of symptoms 20 days or longer before diagnosis and medical treatment). Our early and late postoperative mortality rate was lower than that in medically treated patients.<sup>14</sup>

Dissections of the descending aorta in our series were associated with a mortality rate of 29%. This rate is below that observed by Dalen *et al*<sup>15</sup> and similar to that reported by Miller *et al*<sup>16</sup> in medically treated patients. In the 22 patients with dilatation of the ascending and transverse aorta there were two early and two late deaths. The early and late mortality rates in this group of patients are lower than that in untreated patients.<sup>17,18</sup>

The mortality rate in patients with connective tissue disease who were operated on for dissection or dilatation of the ascending aorta was no higher than the rate in other causal groups; these results are not affected by the inclusion in the group with connective tissue disease of patients aged <40 years in whom the aetiology of aneurysms was described as "unknown" or "other". Thus the risks of operation in patients with Marfan's syndrome do not seem to be increased and their postoperative prognosis in the midterm does not appear to be particularly poor. The implication of this clinical observation is limited by the fairly small numbers of patients we studied.

#### ECHOCARDIOGRAPHY AND CONTRAST ENHANCED COMPUTED TOMOGRAPHY

Echocardiography is the most commonly used non-invasive test for detecting aneurysms of the ascending aorta and serial echocardiographic evaluation is the most practicable method of following up these lesions. Accurate estimation of aortic root dilatation by M mode echocardiography, however, is difficult.<sup>19</sup> Dilatation is often more evident and more pronounced several centimetres above the

valve plane.<sup>20</sup> The lack of correlation between aortic root dilatation and echocardiographic or auscultatory signs of aortic regurgitation may be explained by the difficulty of defining where dilatation starts, especially when it is eccentric, and whether the aortic valve is affected.<sup>21</sup> Cross sectional echocardiography is better than M mode echocardiography for the non-invasive location of aneurysms of the ascending aorta and for determining their extent. The addition of contrast enhanced computed tomography to the diagnostic procedure improved the accuracy of investigation of these lesions in some cases. Preoperative serial echocardiography was not performed in follow up of cases with chronic aortic dissection because dissection requires immediate operation even in symptom free patients. Serial echocardiography had been performed before operation in a follow up of 13 patients in whom aortic root dilatation without dissection had been diagnosed and ten of these also had contrast enhanced computed tomography (Table 7). Contrast enhanced computed tomography was required to confirm the echocardiographic diagnosis when the increased aortic root measurement indicated important progression of disease and in all patients with pronounced aortic root dilatation. In two patients with severe aortic root dilatation (cases 27 and 30) dissection of the ascending aorta was recognised (Fig. 1). In the others echocardiographic diagnoses were confirmed and dissections excluded (Fig. 2). The same diagnoses were reached by contrast enhanced computed tomography and angiography.

In our series cross sectional echocardiography did not detect aortic root dissection in four (21%) out of 19 patients. Because chronic dissection may be missed by echocardiography, we propose that contrast enhanced computed tomography should be performed in patients with aneurysms in whom echocardiography has not been technically satisfactory, in all patients with pronounced aortic root

dilatation or an important increase in dilatation, and in patients with connective tissue disease and symptoms that suggest dissection in the absence of supporting evidence, even those who have only minimal aortic root dilatation. Dissection has been reported in patients with Marfan's syndrome without dilatation of the aortic root.<sup>10</sup>

Postoperatively contrast enhanced computed tomography was used as an outpatient test for evaluating surgical results and for following up distal postoperative dissections. The results of contrast enhanced computed tomography indicated that reoperation should be performed in one case (case 38).

Non-invasive assessment techniques may be of value in identifying patients in whom preventive replacement of the ascending aorta might avoid emergency operation which increases surgical mortality. Donaldson *et al* believe that preventive replacement of the ascending aorta is indicated in symptom free patients with Marfan's syndrome and a large (diameter determined by M mode echocardiography > 60 mm) progressively dilating aortic root or dilated left ventricle (end systolic dimension greater than 55 mm in cases of aortic regurgitation).<sup>22</sup> McDonald *et al* believe that preventive replacement of the ascending aorta should be performed when the aortic root diameter determined by M mode echocardiography is > 55 mm.<sup>23</sup> When the aortic root is 55–60 mm in diameter and the plane of the aortic valve is affected, however, there is almost always severe aortic valve regurgitation which in itself requires replacement of the ascending aorta. Furthermore, there may be severe aortic valve regurgitation when echocardiographic measurement indicates an aortic root diameter of < 55 mm (Table 7). Symptom free patients in whom the aortic root is dilated to > 55 mm without affecting the aortic valve plane and without progressive dilatation of the aortic root do not require preventive replacement of the ascending aorta. In some patients it is difficult to demonstrate a progressive slow dilatation of the aorta by echocardiography alone. When the aortic root is less than 55 mm in diameter but echocardiography and contrast enhanced computed tomography or both have demonstrated a recent rapid progression of dilatation, especially when the aortic valve is affected, prophylactic replacement of the ascending aorta should be considered.

There seems to be no defined limit to aortic root dilatation above which replacement of the ascending aorta should be routinely considered. Even in symptom free patients severe aortic valve regurgitation, the presence of dissection, and the demonstration of rapid dilatation of the aortic root are indications for replacement of the ascending aorta.

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